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A case of Arthrogryposis

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Anesthetic Management in a Patient with Arthrogryposis

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INTRODUCTION

Arthrogryposis is a spectrum of disorders characterized by persistent multiple limb contractures. It is often associated with myopathies, spinal muscular atrophy, and hypotonia.

Due to the abnormal craniofacial features along with severe limb and neck contractures, patients with Arthrogryposis syndromes often present with difficult airway management, intravenous (IV) line placement, neuraxial technique and patient positioning.

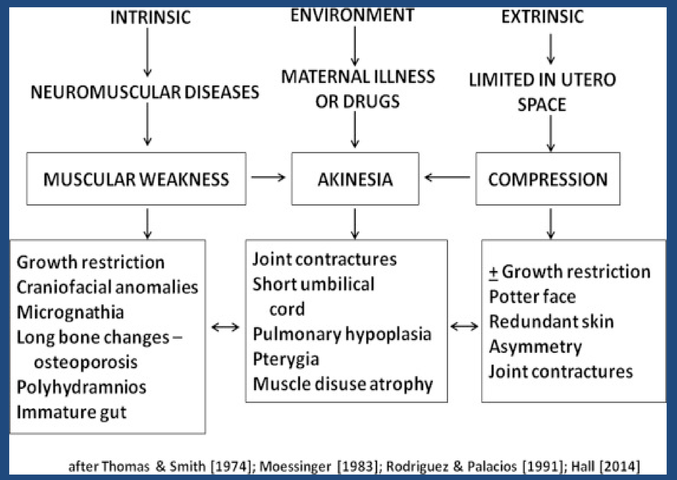
CASE DESCRIPTION

A 9-year-old female with history of congenital bilateral talipes equinovarus with underlying Arthrogryposis syndrome status post triple arthrodesis presented for bilateral cast removal and short leg cast application.

The patient received inhalation masked induction with sevoflurane and nitrous oxide. A Laryngeal Mask Airway (LMA) was placed after successful placement of left hand peripheral IV. General anesthesia was maintained with sevoflurane inhalation. Patient's temperature was monitored closely during the intraoperative period.

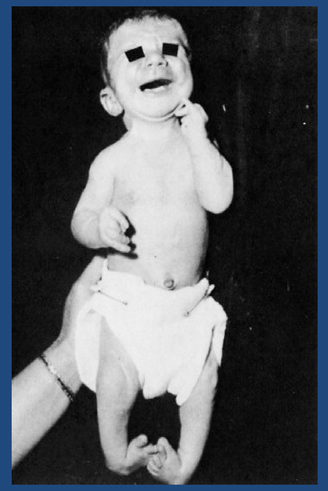
Her perioperative course was uneventful and patient was discharged home same day of surgery.

Figure 1: Mechanism of Secondary Effects of Fetal Akinesia[5]



Hall, J. Arthrogryposis (multiple congenital contractures): Diagnostic approach to etiology, classification, genetics, and general principles. *European Journal of Medical Genetics*. July 2014 . Image obtained from Clinical key.

Figure 2. Distal Arthrogryposis Type I[6]
Note predominant distal contractures, with overlapping finger contractures, ulnar deviation, and clubfeet.



Hall, J. et al. Emery and Rimoin's Principles and Practice of Medical Genetics. Arthrogryposes (Multiple Congenital Contractures). Figure 161-2. Jan 2013. Image obtained from Clinical Key.

Table 1. Precautions before Anesthesia and Anesthetic Considerations[1-6]

Risk of Difficult Airway	-Micrognathia -Limited temporomandibular joints mobility -Atlanto-occipital instability -Fusion of cervical vertebrae -Short neck -Craniofacial anomalies
Difficult Venous Access and regional block	-Severe flexion/contracture abnormalities of the extremities -Joints pterygia
Abnormal cardiopulmonary function	-Severe scoliosis -Congenital cardiomyopathy -Congenital hypoplastic lungs, tracheal stenosis
Neuromuscular Disease Risk of hyperkalemic response to succinylcholine Risk of aspiration	-Muscle atrophy -Absence of muscles -Muscle structural abnormalities
Risk of hyperthermia	-Hypermetabolism

DISCUSSION

Most Arthrogryposis Syndrome cases are of sporadic inheritance.
Incidence varies from 0.3-3 cases per 1000 live births.[1]

- Arthrogryposis Multiplex Congenita (AMC), most severe arthrogryposis syndrome, are characterized by multiple joint contractures with coexisting comorbid multisystem disease such as pharyngeal, cardiac, urologic, and gastric abnormalities.[2]
- The less severe, Distal Arthrogryposis Syndromes (DAS) only involve distal joints such as hands and feet. Neurologic development is normal and response to physical therapy is usually good in patients with DAS.

Clinical diagnosis is usually made at birth. Severe equinovarus deformity of the feet usually presents. The face is usually round, flat nasal bridge, and micrognathia. Contractures and rigidity of the joints are common and result in joint pterygia and dislocation of large joints (hip, knee, shoulder and elbow). There is significant decrease mobility of temporomandibular joint, atlantoaxial instability, scoliosis, and/or fusions of cervical vertebrae have been reported.[1]

Abnormal craniofacial features along with severe limb and neck contractures result in difficult airway management, therefore spontaneous ventilation is recommended until the airway is secured. The use of LMAs, fiberoptic intubation, or fiberoptic-guided intubation via LMAs have been proposed and used successfully. Careful intraoperative positioning is mandatory to prevent injuries and fractures. Risk of hypermetabolism and hyperpyrexia during and after anesthesia has been described in prior case reports. However, no relation between malignant hyperthermia and arthrogryposis has been demonstrated.[1-3] An exaggerated hyperkalemic response to succinylcholine may occur secondary to muscle structure changes.

CONCLUSION

Patients with Arthrogryposis syndromes are at increased risk of difficult airway management, challenging vascular access, and hypermetabolic state with hyperthermia in the setting of myopathies. Along with joint contracture problems and associated congenital anomalies (cardiac, respiratory, genitourinary, neurogenic, and facial anomalies) present further challenges for anesthesiologists.
Therefore, careful airway evaluation, close perioperative monitoring of vitals for hyperthermia, tachycardia, and hypercarbia, and preparation for anticipated emergencies are needed to avoid critical events.

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